

# Frontotemporal Dementia

**American Academy of  
Neurology**

**John Hart, Jr., M.D.**

# Frontotemporal Dementia

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- **Definition: clinicopathologic condition consisting of deterioration of personality and cognition assoc. with prominent frontal and temporal lobe atrophy**
- **Accounts for up to 3-20% of dementias**
  - **Third behind AD and Lewy Body Dementia in neurodegenerative dementing illnesses**

# Prevalence

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- **Mean age of onset 52.8** (Ratnavalli et al. Neurology 2002;58:1615-1621)
- **Male preponderance 14:3 in one study and M=F in others**
- **Dementia prevalence of 81 per 100,000 (95% CI, 62.8 to 104.5) in the 45-64 year age group**
- **Prevalence of AD and FTD in 45-64 age group same at 15 per 100,000 (8.4-27.0)**

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- Thus, common cause of dementia in younger population
- Greater caregiver burden and increased dependency and health care costs

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- **Established clinical consensus criteria** (The Lund and Manchester Groups, J Neurol Neurosurg Psychiatry 1994;57:416-418; Neary et. al, Neurology 1998;51:1546-1554):
- **Core features**
  - **Insidious onset and slow progression**
  - **Early decline of**
    - **Social interpersonal conduct**
    - **Regulation of personal conduct**
    - **Insight**
  - **Early emotional blunting**

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- **Supportive features:**
  - **Decline in personal hygiene and grooming**
  - **Mental rigidity and inflexibility**
  - **Distractibility and impersistence**
  - **Hyperorality**
  - **Perseverative behavior**
  - **Speech and language**

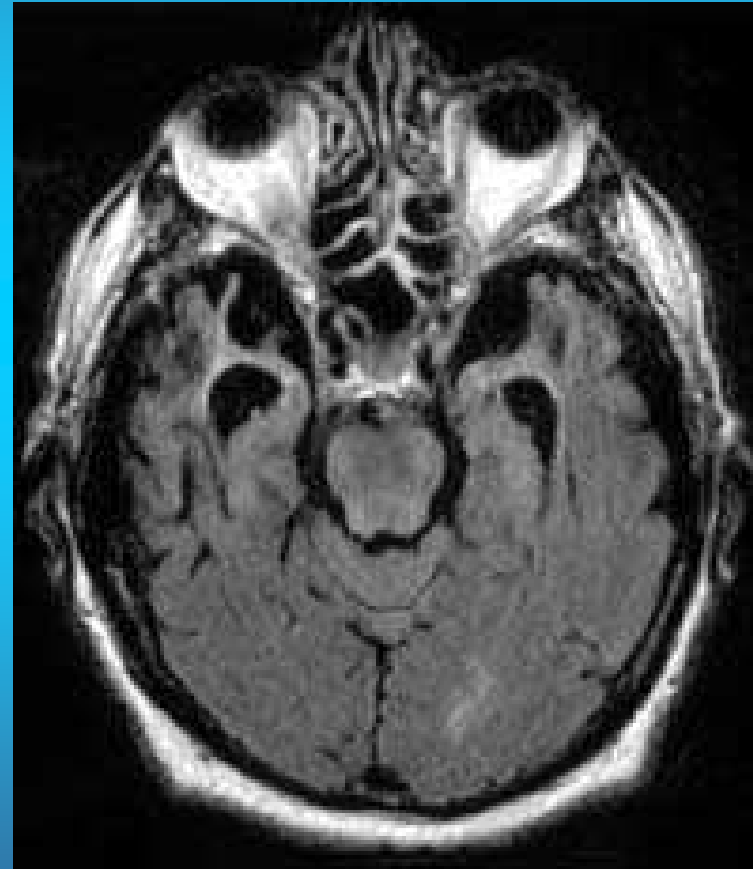
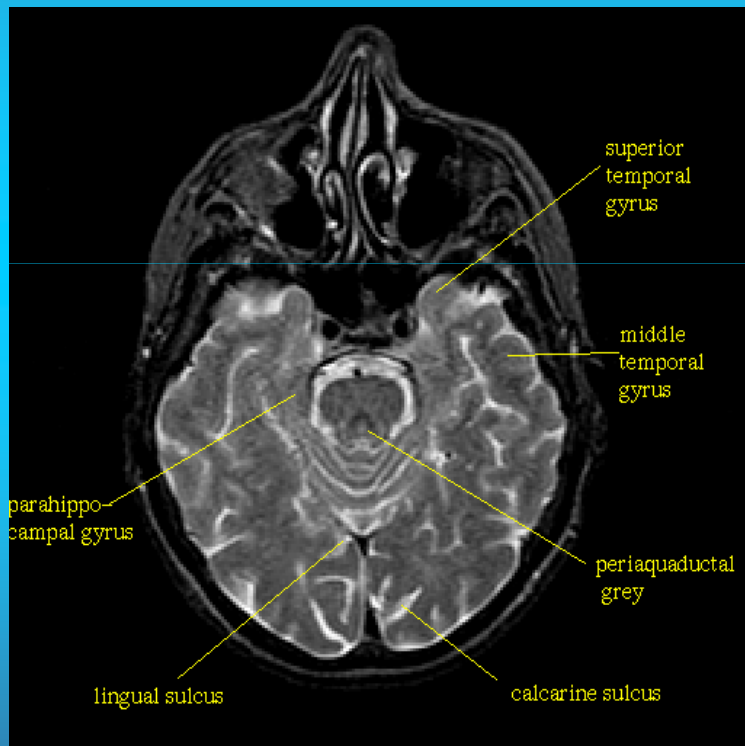
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- Neuropsychology:
  - Impaired frontal lobe tests in absence of severe amnesia, aphasia, or visuospatial deficits
- Imaging:
  - Atrophy or decreased uptake in the frontal or anterior temporal lobes (bilateral or unilateral) by MRI, CT, PET, SPECT (The Lund and Manchester Groups, J Neurol Neurosurg Psychiatry 1994;57:416-418; Neary et. al, Neurology 1998;51:1546-1554)

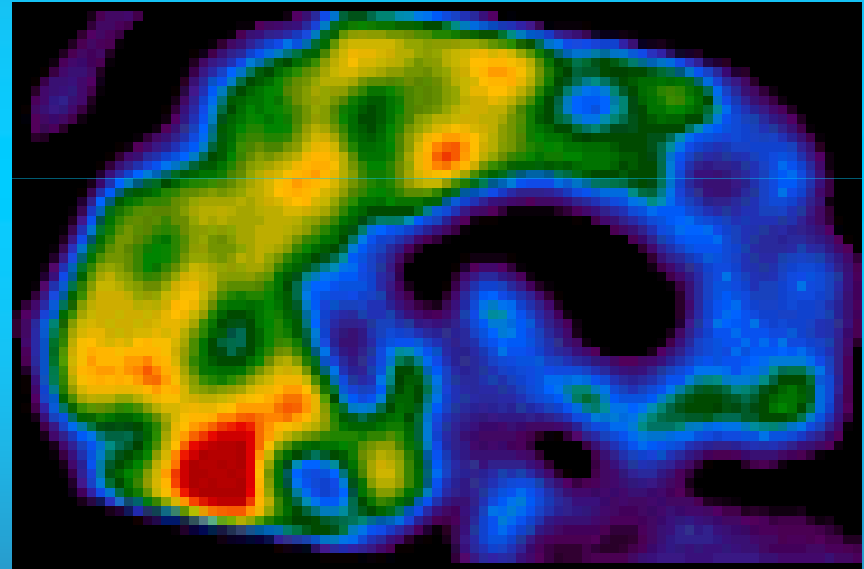
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- Recent study showed of 42 FTD cases, 19 had at least 1 other family member affected (Chow et al., Arch Neurol. 1999;56:817-822)
  - 1/3 had a positive family history in this study, but others much lower
- Appears to be inherited in autosomal dominant fashion in those patients
- Linked to chromosome 17 (tau gene) and chromosome 3
- Association with motor neuron disease (ALS)

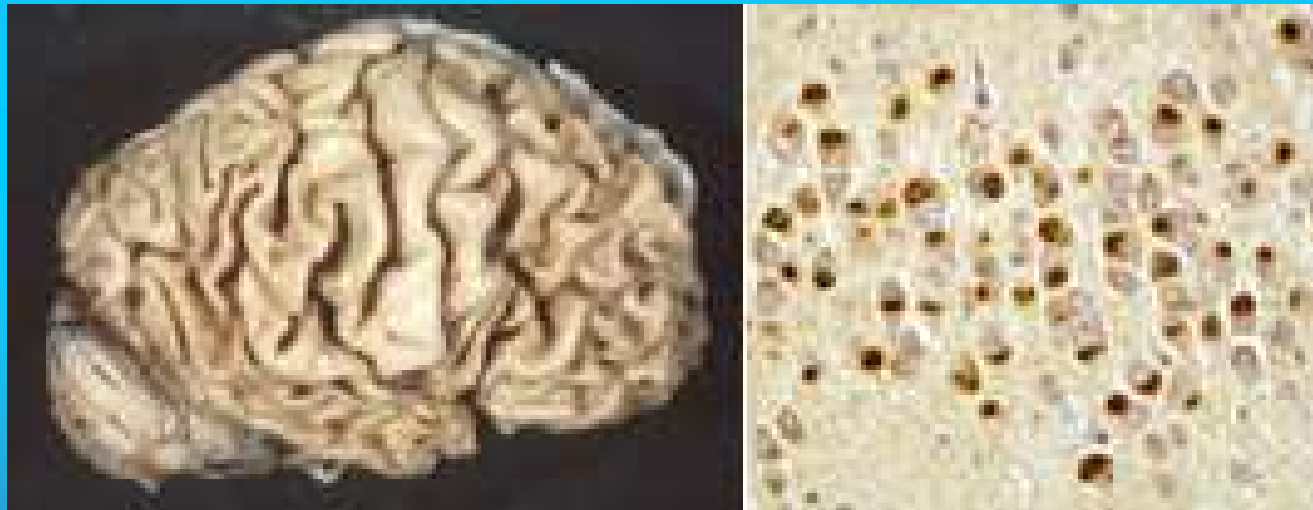
# Pathology

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- **Prominent frontal and temporal lobar atrophy**
- **Atrophy may be associated with Pick's bodies, tauopathy, nonspecific superficial cortical neuron loss (DLBDH)**

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- Term is conventionally used in clinical and pathological diagnoses in standard clinical setting
- No other specific code comes reasonably close to capturing disease
- Dementia or AD diagnosis (290 or 331) ignores established diagnostic criteria and allows no tracking of FTD

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- Differs from the other codes including AD, frontal dementia, Pick's disease
  - Use of these codes inappropriate as not capture the age of onset, duration of illness, genetic factors, and impact on caregiver, society, and economics
- AD → older, different duration, less clear genetics
- Frontal dementia → no temporal lobe involvement, genetics differ
- Pick's disease → not capture spectrum of FTD

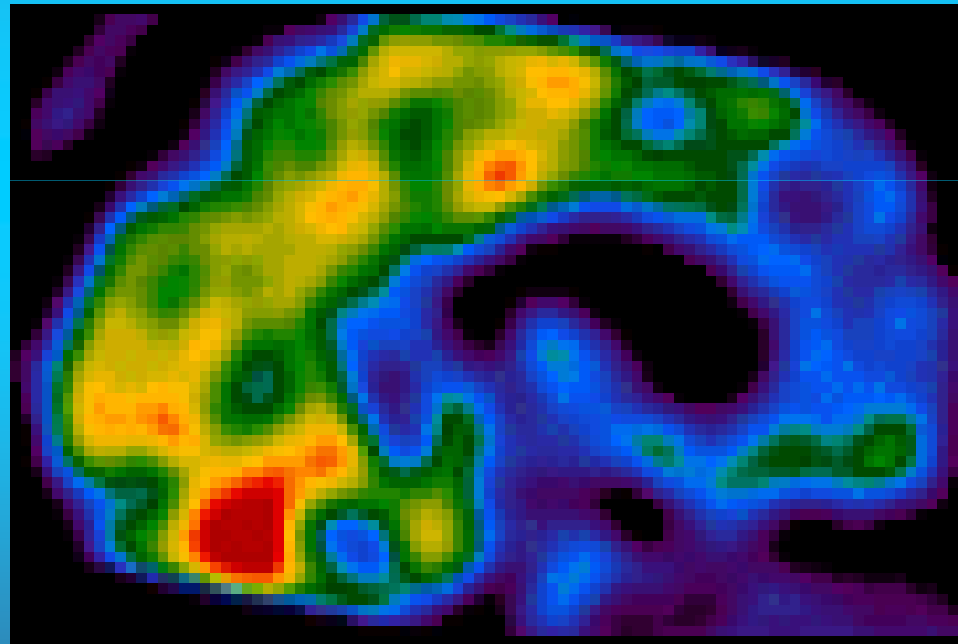
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- **Implications for FTD different from other dementias/AD:**
  - **Greater caregiver burden and increased dependency and health care costs**
  - **Patients see codes and think they have some other disease**

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